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Abstract: Objective: Diaphragmatic paralysis (DP) caused by phrenic nerve injury is potentially life-threatening in infants. Phrenic nerve injury due to thoracic surgery is the most common cause of DP in children. We retrospectively analyzed incidence, surgical details, management and follow-up of our patients with DP after cardiac surgery to develop an algorithm for the management and follow-up. Methods: Retrospective analysis of 43 patients with DP after cardiac surgery performed between 1996 and 2000. Results: Median age at cardiac surgery was 1 month (range 3 days to 9 years). Incidence of DP was 5.4%. A trend towards higher incidences of DP were observed after arterial switch operation (10.8%, $P=0.18$), Fontan procedure (17.6%, $P=0.056$) and Blalock-Taussig Shunt (12.8%, $P=0.10$). Median time from cardiac surgery to surgical plication was 21 days (range 7-210 days). Transthoracic diaphragmatic plication was performed in 29/43 patients, no plication was done in 14/43 patients. Patients in whom diaphragmatic plication was required were younger (median age 2 months, range 21 days to 53 months versus 17.5 months, range 4 days to 110 months; $P 0.001$). Indications for plication were failure to wean from ventilator ($n=22$), respiratory distress ($n=4$), cavopulmonary anastomosis ($n=2$), and failure to thrive ($n=1$). All these symptoms resolved after diaphragmatic plication, however, 8/29 patients with plication and 2/14 without plication died. Cause of death was not related to diaphragmatic plication in any patient. Position of plicated diaphragm was normal in 18/21 surviving patients 1 month after plication. In 2/12 surviving patients without plication hemidiaphragm showed a normal position 1 year after surgery. The rate of pulmonary infections was not significantly different during 12-60 months follow-up. Conclusions: DP is an occasional complication of cardiac surgery. High incidences of DP were seen after arterial switch operation, Fontan procedure and Blalock-Taussig shunt (BT). Respiratory insufficiency requires diaphragmatic plication in most infants with DP whereas older children may tolerate DP. Transthoracic diaphragmatic plication is an effective treatment of DP and achieves relief of respiratory insufficiency in most patients. Spontaneous recovery from postsurgical DP is rare

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Incidence and treatment of diaphragmatic paralysis after cardiac surgery in children

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Abstract

Objective: Diaphragmatic paralysis (DP) caused by phrenic nerve injury is potentially life-threatening in infants. Phrenic nerve injury due to thoracic surgery is the most common cause of DP in children. We retrospectively analyzed incidence, surgical details, management and follow-up of our patients with DP after cardiac surgery to develop an algorithm for the management and follow-up. **Methods:** Retrospective analysis of 43 patients with DP after cardiac surgery performed between 1996 and 2000. **Results:** Median age at cardiac surgery was 1 month (range 3 days to 9 years). Incidence of DP was 5.4%. A trend towards higher incidences of DP were observed after arterial switch operation (10.8%, $P=0.18$), Fontan procedure (17.6%, $P=0.056$) and Blalock-Taussig Shunt (12.8%, $P=0.10$). Median time from cardiac surgery to surgical plication was 21 days (range 7-210 days). Transthoracic diaphragmatic plication was performed in 29/43 patients, no plication was done in 14/43 patients. Patients in whom diaphragmatic plication was required were younger (median age 2 months, range 21 days to 53 months versus 17.5 months, range 4 days to 110 months; $P<0.001$). Indications for plication were failure to wean from ventilator ($n=22$), respiratory distress ($n=4$), cavopulmonary anastomosis ($n=2$), and failure to thrive ($n=1$). All these symptoms resolved after diaphragmatic plication, however, 8/29 patients with plication and 2/14 without plication died. Cause of death was not related to diaphragmatic plication in any patient. Position of plicated diaphragm was normal in 18/21 surviving patients 1 month after plication. In 2/12 surviving patients without plication hemidiaphragm showed a normal position 1 year after surgery. The rate of pulmonary infections was not significantly different during 12-60 months follow-up. **Conclusions:** DP is an occasional complication of cardiac surgery. High incidences of DP were seen after arterial switch operation, Fontan procedure and Blalock-Taussig shunt (BT). Respiratory insufficiency requires diaphragmatic plication in most infants with DP whereas older children may tolerate DP. Transthoracic diaphragmatic plication is an effective treatment of DP and achieves relief of respiratory insufficiency in most patients. Spontaneous recovery from postsurgical DP is rare.

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Keywords: Diaphragmatic plication; Phrenic nerve injury; Infant; Newborn

1. Introduction

Diaphragmatic paralysis (DP) due to phrenic nerve injury is a rare respiratory condition which may be life-threatening in infants and young children [1-3]. Prior to the advent of cardiac surgery for congenital heart disease, most phrenic nerve injuries and DP were secondary to birth trauma. In recent years an increasing number of phrenic nerve injuries associated with thoracic surgical procedures has been reported [4-9]. Today, it is the most common cause of DP in children with an incidence of 0.3-12.8% [3,5,10,11].

DP may present with respiratory distress, atelectasis, recurrent pneumonia or inability in weaning from ventilator.

In contrast to infants and younger children, older children can compensate the loss of diaphragmatic function and usually present with little or no symptoms [3,11-17].

The diagnosis of DP is difficult and can be easily missed in older children. DP may be suspected by elevated hemidiaphragm on chest X-rays, however, the confirmation requires diaphragm mobility tests by ultrasound and/or fluoroscopy during spontaneous breathing.

During the last years, diaphragmatic plication has evolved as surgical treatment for DP in patients with respiratory distress or inability to wean from ventilator. In infants and children under 1 year of age diaphragmatic plication has become a standard treatment for DP. However, little is known about the long-term follow-up of children with surgical DP and after diaphragmatic plication.

The aims of this retrospective study were to assess the incidence of DP after cardiac surgery, potential risk factors, and to describe the management and long-term follow-up.

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2. Patients and methods

2.1. Patients

Between January 1996 and December 2000, 802 consecutive pediatric patients underwent cardiac surgery for congenital heart disease. Postoperative DP was diagnosed in 43/802 (5.4%) patients. Personal data and surgical notes were collected by chart review. Follow-up data were obtained by a family physician/pediatrician questionnaire and cardiology outpatient follow-up notes. In addition, chest X-rays within 1 month and 1 year after plication, or 1 year after cardiac surgery if the diaphragm was not plicated, were reviewed.

2.2. Diagnosis

A diagnosis of DP was suspected by failure to wean from assisted ventilation, elevation of the diaphragm on chest X-ray, or paradoxical movement of the epigastrium during spontaneous ventilation. DP was confirmed in all cases with ultrasound and/or fluoroscopy during spontaneous breathing. All patients had pre-operatively an X-ray and none of them showed sign of diaphragmatic paralysis like a higher standing diaphragm before operation.

2.3. Treatment

Indications for diaphragmatic plication were aged under 6 months, respiratory distress and the inability to wean from ventilator and children with cavopulmonary shunts with the intention to avoid an increase in pulmonary vascular resistance. Respiratory distress was defined as one or more of the following criteria: tachypnoea, oxygen dependency or CO₂ retention. Diaphragmatic plications were done using a thoracic approach as described by Bisgard [22]. In all patients with unilateral paresis the plication was performed through the seventh intercostal space with a lateral thoracotomy and fixation of the diaphragm on the ventral 10th costal arch with non-absorbable braided sutures, sometimes with pledges. In the three cases with bilateral paresis only one side was plicated similarly.

2.4. Statistics

Descriptive statistics are reported as median values and range. Student's *t*-test were used for comparison between groups. Two analysis were applied for determination of risk factors for DP. A *P* value of <0.05 was considered statistically significant.

3. Results

The incidence of DP was 5.2% after open-heart surgery and 5.8% after closed-heart surgery. Fig. 1 depicts the age distribution with 77% infants being under 1 year and 37% under 1 month of age at the time of cardiac surgery.

Type and distribution of cardiac surgery causing DP is depicted in Table 1. A trend toward a higher incidence of DP was seen after Fontan procedures (17.6%; *P*=0.056), BT

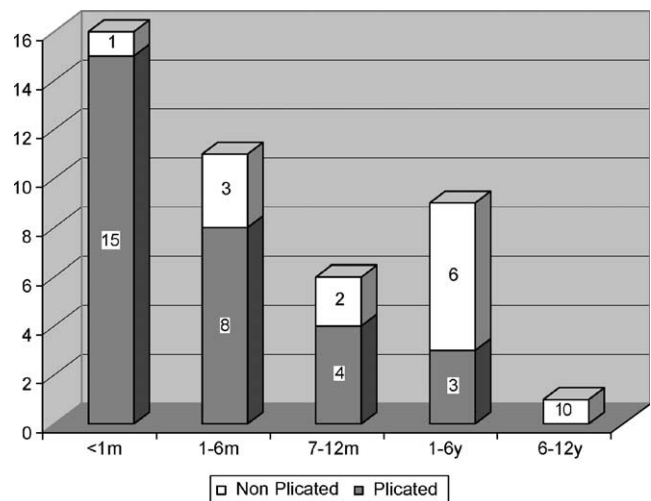


Fig. 1. Distribution of age in plicated and non-plicated patients.

Shunt (12.8%; *P*=0.10), arterial switch operation (10.8%; *P*=0.18; Table 2).

In 21/24 (88%) intubated patients DP was first suspected due to difficulties or inability to wean from ventilator. One patient showed an elevated hemidiaphragm on chest X-ray, one was coincidentally found during an abdominal ultrasound, and one patient showed an asymmetric breathing pattern during weaning.

In 9/19 (47%) extubated children, DP was first suspected by respiratory distress, an asymmetric breathing pattern in 4/19 (21%), elevation of the diaphragm on a routine chest X-ray in 4/19, failure to thrive or a routine abdominal ultrasound in one patient each.

Table 1
Numbers of the different surgical procedures resulting in DP

	Previous surgery	No previous surgery	Total	%
Arterial switch operation	0	5	5	11.6
Ventricular septal defect closure	0	4	4	9.3
Fontan operation	3	0	3	7.0
Pulmonary artery unifocalisation	2	1	3	7.0
Repair of atrioventricular septal defect	0	2	2	4.7
Central aortopulmonary shunt	2	0	2	4.7
Miscellaneous	6	5	11	25.6
Subtotal	13	17	30	69.8
Closed-heart procedures				
Blalock-Taussig shunt	0	5	5	11.6
Repair of coarctation	0	3	3	7.0
Repair of coarctation and pulmonary artery banding	0	2	2	4.7
Pulmonary artery banding	1	1	2	4.7
Pexie of descending aorta	1	0	1	2.3
Subtotal	2	11	13	30.2
Total	15	28	43	100.0

Miscellaneous: anastomosis collector vessel-left atrium (1); hemi Fontan (1); Glenn shunt (1); repair total anomalous pulmonary venous drainage (1); pulmonary artery debanding (1); pulmonary artery reduction (1); repair of fallot (1); repair of truncus arteriosus (1); mitral valve reconstruction (1); widening of a pulmonary vein collector vessel (1); repair of DORV (1).

Table 2
DP incidence after specific surgeries

	VSD closure	Switch	Fontan	BT shunt
1996-2000	85	46	17	39
DP cases	4	5	3	5
Incidence (%)	4.7	10.8	17.6	12.8

Confirmation of DP was achieved by ultrasound alone in 19/43 (44%), by fluoroscopy alone in 13/43 (30%), and by both techniques in 11/43 (26%) cases.

The paralysed hemidiaphragm was left sided in 23/43 (53.5%), right sided in 17/43 (39.5%) and bilateral in 3/43 (7%) cases.

The assisted ventilation time after cardiac surgery was for patients with plication 13 days (range 1-54 days) and for patients without plication 5 days (range 1-49 days). The median time from cardiac surgery to surgical plication was 21 days (range 7-210 days). Median age of patients who required plication was 2 months (range 21 days to 53 months) and from patients without plication 17.5 months (range 4 days to 110 months; $P < 0.001$). Of 10/43 (23%) patients with DP who died, eight required diaphragmatic plication. Deaths were due to different causes, but not due to the plication procedure (Table 3). None of these patients had a bilateral DP. The three patients with bilateral DP required plication only on the left-side.

One month after plication, plicated diaphragm position was normal on chest X-ray in 18/21 (86%) plicated survivors. One year after plication, the affected diaphragm remained in a normal position in 13/21 (62%) cases. In 3/21 (14%) it was elevated due to atelectasis, and in 2/21 (19%) X-rays were not available. Three plicated survivors (14%) showed an elevated diaphragm 1 month after plication. In one of these patients diaphragm returned to a normal position, another remained elevated and the third had no X-ray follow-up. One year after cardiac surgery, just 2/12 non-plicated surviving patients showed a normally positioned hemidiaphragm.

The median number of pulmonary infections in both plicated and non-plicated patients were 1.5 (range 1-4 infections per year) per year.

4. Discussion

Diaphragmatic paralysis (DP) remains a relatively rare respiratory condition which may be life-threatening in infants and young children. In our retrospective analyses

the 5.4% incidence of DP after cardiac surgery is comparable to other retrospective studies that showed an incidence of 0.3-5.7% [3,8-10]. In prospective studies the reported incidence varies from 0.5 up to 12.8% which may indicate a substantial number of undiagnosed patients or difficulties in diagnosis [3,9,11,12].

The age of plicated patients was significantly lower compared to the non-plicated. This confirms the observation that infants and younger children tolerate DP less than older children because infants depend mainly on diaphragmatic contraction for adequate gas exchange. The intercostal muscles are weaker and there is a more horizontal orientation of the rib cage. In addition, infants have an increased mediastinal mobility with shifting of mediastinal contents to the contra lateral side on inspiration with DP while the paralysed diaphragm is pulled upwards. On the ipsilateral side, the diaphragm cannot resist negative intrapleural pressure and moves paradoxically. This reduces functional residual capacity, facilitates alveolar collapse and atelectasis. Moreover, the infants recumbent position leads to a reduction of vital capacity and their small intrabronchial calibres facilitates obstruction and atelectasis by retained secretions [3,11,13-17]. The highest incidence of DP after open-heart surgery was seen after Fontan operation. This might be due to the surgical technique using extracardiac conduits which requires a more extensive thoracic preparation than other operations.

The high incidence of DP after arterial switch operations has been reported in the literature [3,11,14,18,19]. A reason may be that arterial switch operations often require harvesting of autologous pericardium and/or thymus resection [3,9-11,13,14,16,18]. All cardiac surgery on bypass were done in mild to moderate hypothermia depending from complexity and duration of surgery.

In the group of patients who underwent closed-heart procedures Blalock-Taussig shunts most often caused DP. Our results with an incidence of 11.6% are similar to other series with an incidence of 2.1-19% after Blalock-Taussig shunts [3,9-11,13,14,16-18]. Another factor increasing the risk of DP is previous thoracic surgery. In our series 35% of the patients underwent previous surgery. The reported rate in the literature varies also from 9 to 49% [3,9,11,13,14].

Surgical plication is today the widely accepted treatment of DP especially in children under 1 year of age. However, there is still controversy on its best timing. Some authors recommend that plication should be performed as soon as the diagnosis of DP has been confirmed [6,7] while others recommend a waiting period of 1-6 weeks in anticipation of potential spontaneous recovery [5,12-14,16-18].

We agree with other authors who suggest the decision of plication should be based on the respiratory status of the patient [3]. In our series, median time from cardiac surgery to surgical plication was 21 days (range 7-210 days). This considerable time period was in most cases due to the critical clinical status of the patient. We could not observe any case of spontaneous recovery in the first 4 weeks after heart surgery.

In earlier years the use of mechanical ventilation was favored as the treatment of choice. Haller et al. suggested a trial of continuous positive airway pressure (CPAP) during 4-6 weeks [2,20]. This time should allow to

Table 3
Diagnosis of the patients who died

Pulmonary hypertension	1
Myocardial infarction due to coronary stenosis	1
Arrhythmia (not plicated)	1
Tracheal stenosis	1
Liver failure + spasticity of upper resp. tract	1
T cell defect with CD4 lymphopenia	1
Unclear dysmorphic syndrome	1
Trisomie 18	1
Unclear tetraparesia (not plicated)	1
Recurrent pneumonia and insufficient plication	1
Total	10

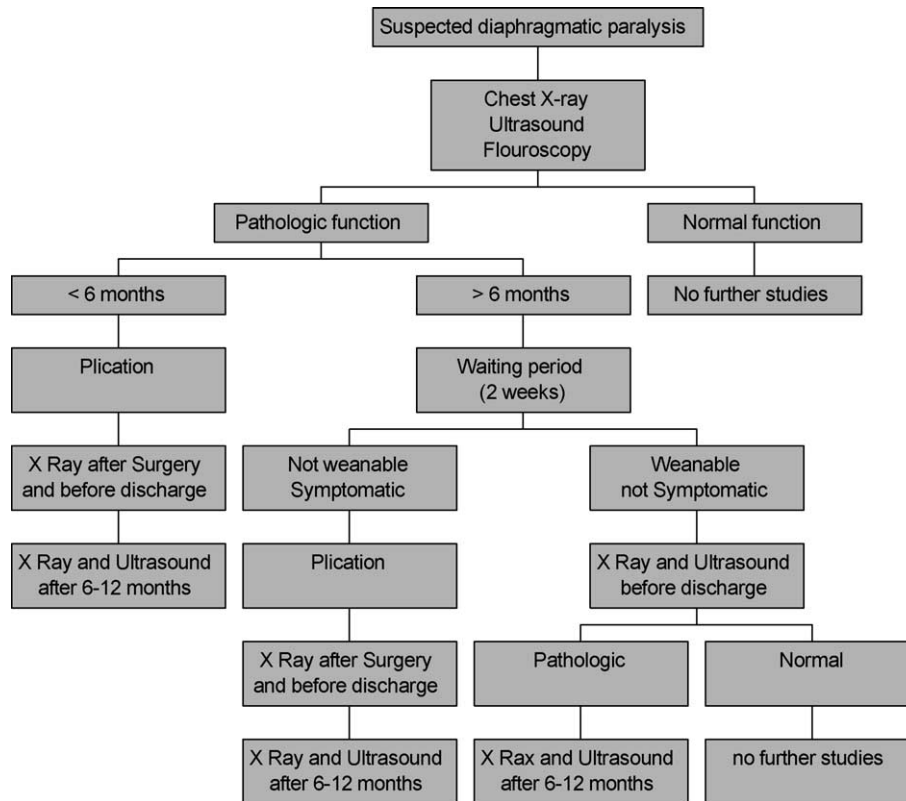


Fig. 2. Algorithm of patients with diaphragmatic paralysis.

differentiate children with respiratory dysfunction from children who will benefit from plication. However, late surgical plication may be jeopardized by atrophy of the diaphragm which may even preclude successful surgical plication.

Regarding the impact of plication on ventilation time and hospital stay, there are reports which describe a reduction after plication [8-11,15]. In our study we analyzed the patients retrospectively. For this reason the groups of plicated and non-plicated patients are not exactly comparable. Although surgical plication of DP improved symptoms in most patients the mortality of patients with DP remained high, due to complex congenital heart disease and complex cardiac surgery.

To assess the function of the diaphragm after DP in plicated and non-plicated patients the diaphragmatic position on X-rays and the frequency of pulmonary infections were followed. Using the definition of Greene et al. [4], 86% of surviving plicated patients showed a diaphragm in a normal position 1 month after plication. The return to normal position after plication is also documented in the literature [5-8,11].

Pre-disposition to pulmonary infections and pneumonia is a well known clinical sign of DP. Using the definitions of Feigin et al. [21] that it is 'normal' in infancy to have 3-8 respiratory infections (with no hospital admission required) per year, none of our groups showed a higher pre-disposition to respiratory infections (1.5 infections per year).

Ciccollella et al. [23] explained in a paper the physiological success of plication. During inspiration the healthy

diaphragm produced negative intrathoracic pressure and the abdominal contents is drawn into the paralyzed side of the thorax. This paradoxical motion does not expand the lung on this side and results in poor gas exchange. After plication the paralyzed side is more resistant again this pressure and the adjacent lung segments expand [23]. The non-plicated patients were mostly older than 12 months so they have better compensatory mechanisms to cope with DP.

Prospective studies need to be done to evaluate diaphragmatic function of the plicated and non-plicated patients including pulmonary function tests at long-term follow-up. Based on our results, we suggest an algorithm for the study of patients with DP (Fig. 2).

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